THE OPERATIVE TREATMENT OF HYDROCEPHALUS: A PRELIMINARY REPORT OF FORTY-ONE PATIENTS.[1]

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The treatment of hydrocephalus has been most discouraging. One operative method after another has been advocated as a possible means of successful drainage in these patients; one or more cases are reported and then nothing more is heard of the method, the patients having died and the method no longer used. Frequently a "new" method of drainage is published more upon a theoretical basis rather than upon practical applicability; one case alone is published as having been operated upon and the result was death; surely, we must grant that the method may have been new, but the result has been the same as in the earlier methods of treatment, and unless the percentage of cures, or even improvement, can be raised, such methods are naturally soon discarded.

The chief cause of failure in these methods of drainage is, in my opinion, not due so much to the ignorance regarding the nature of hydrocephalus itself, what the cerebrospinal fluid really is, its method of secretion and excretion and similar points of almost total obscurity, but rather to a lack of appreciation of the fact that the condition of hydrocephalus is rarely limited to a dilatation of the ventricles alone—that is, the so-called hydrocephalus interna, which is the result of a blockage of the escape of cerebrospinal fluid from the ventricles into the subarachnoid spaces by an obstruction of the adequate of Sylvius or of the foramina of Majendie and Luschka; but, on the contrary, the condition is most frequently due to a lessened excretion of the cerebrospinal fluid through the subarachnoid cranial and spinal veins, the sinuses, possibly lymphatics, etc., so that the type of so-called hydrocephalus externa is developed. It is this latter condition of so-called hydrocephalus externa that is most frequent and yet the methods of treatment have been almost all of them directed to a drainage of the ventricles in the condition of hydrocephalus when it is a fact that the ventricles do not require drainage in only a small percentage of these cases; they are not dilated in the majority of cases because the aqueduct of Sylvius and the foramina of Majendie and Luschka are not obstructed, and there is a free escape of cerebrospinal fluid from the ventricles into the subarachnoid spaces, so that any operation merely directed to connect the ventricles with the subarachnoid or subdural spaces in those cases is of no possible benefit. In the comparatively rare cases, however, of true hydrocephalus interna due to a definite blockage of the aqueduct of Sylvius or of the foramina of Majendie and Luschka by a subtentorial lesion, those methods of drainage are logical, and yet I believe they are of very limited usefulness in that the condition of hydrocephalus interna is almost always, in my opinion, the result of an early diffuse meningitis, the results of which (adhesions, exudate, thickenings, etc.) may not only block the escape of cerebrospinal fluid from the ventricles into the subarachnoid spaces, but also so impair and even completely block the channels of escape of the cerebrospinal fluid through the cortical veins, sinuses, possibly lymphatics, etc., so that even if the ventricles were connected with the subarachnoid spaces, yet the cerebrospinal fluid would not be drained from the cerebrospinal canal; that is, the condition of blockage of the escape of cerebrospinal fluid is a diffuse one rather than a mere obstruction of the ventricles; there are cases, to be sure, of hydrocephalus interna due to a tumor or cyst formation blocking the aqueduct of Sylvius or the foramina of Majendie and Luschka, and where the ventricular subarachnoid method of drainage would be logically applicable, but these cases form but a very small percentage of the large group of hydrocephalus patients; in this series of forty-one patients, twenty-six were of the external type, whereas only fifteen were of the internal type of hydrocephalus; besides, the method of draining the cerebrospinal fluid described in this paper can be used in either condition—that is, whether the condition is one of the internal or of the external type of hydrocephalus. The condition of unilateral hydrocephalus may be mentioned as a very rare occurrence; it is due to a blockage of the aqueduct of Sylvius and the opposite foramen of Monro; one of our patients was of this type.

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[1] A complete report of the cases in detail will be published later.

In order to differentiate whether the condition is one of hydrocephalus interna or of externa, or of both (and this last condition occurs fairly frequently), the method of Dandy and Blackfan[2] in their excellent monograph can be used whereby the time of excretion of phenolsulphonaphthalein first from the ventricles and then from the spinal subarachnoid spaces can be observed and the amount estimated in the urine; if practically of equal duration and amount then the ventricles are not blocked and the condition is one of hydrocephalus externa, or of both, whereas, if the excretion of phenolsulphonephthalein injected into the ventricles is greatly delayed and lessened in amount in the urine, then the ventricles are blocked and the condition is one of hydrocephalus interna. A much more simple method, however, can be used, and I believe it is of sufficient accuracy to warrant its use whenever a differentiation of the two conditions is desired in the treatment of hydrocephalus. I make this statement because the operative method to be described in this paper makes such a preliminary test unnecessary, as the differentiation of the internal from the external type of hydrocephalus can be ascertained at the time of the operative procedure and treated accordingly through the same operative incision and at the same time. If, however, it is desired to ascertain the type of hydrocephalus before the operation, then a ventricular puncture needle can be inserted into the ventricle through a very small skin incision at the anterior fontanelle as far from the median line as possible, and at the same time a spinal puncture needle is inserted into the lumbar subarachnoid space; with the patient lying upon his side, the median line of the head being upon an exact level with the spinal canal and the patient being perfectly quiet, the pressure and rate of flow of the cerebrospinal fluid from both the needles should be the same if the ventricles and subarachnoid spaces are in free communication; the condition would therefore be one of hydrocephalus externa. If, however, the ventricles are blocked, then the pressure and rate of flow of cerebrospinal fluid from the ventricle needle is greatly in excess of that from the spinal puncture needle, which may be but a few drops; the condition therefore would be one of hydrocephalus interna. Another aid in the differentiation in the older children may be found in the roentgen-ray—the internal type producing convolutional markings throughout the vault of the skull, whereas this would naturally not occur in hydrocephalus externa.

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[2] Am. Jour. Dis. Child., December, 1914, pp. 406–482.

Many operative procedures have been used in the past to combat the condition of hydrocephalus. In the early days of Hippocrates merely repeated lumbar punctures and ventricular punctures were performed, but the result was invariably the same—a steady advance of the condition and frequently a much more rapid progress following the tapping, as though the sudden removal of the increased amount of the cerebrospinal fluid stimulated in some way a greater secretion of the cerebrospinal fluid so that the condition rapidly became worse. Occasionally, however, a case has been reported improved by such treatment; such cases, however, were rare and the reports of them were seldom made after one year had elapsed; they may have been, therefore, merely temporarily benefited. Drainage of the cerebrospinal fluid externally to the skin has almost always resulted fatally on account of the consequent infection and the resulting meningitis.

For these reasons many complicated and most tremendous operations have been devised to drain successfully the cerebrospinal fluid from the ventricles and the subarachnoid spaces, and all of them have been most disappointing.

Draining the cisterna magna (fourth ventricle) into the adjacent sinuses or into the pleural cavity by means of strands of various material through a suboccipital opening; connecting the spinal subarachnoid canal with the retroperitoneal space by means of a laminectomy and a laparotomy (and naturally limited to cases of external hydrocephalus); joining the third ventricle to the subarachnoid spaces by means of a tube through the corpus callosum and the various other methods described in the literature—usually these methods have been abandoned after a few patients have been operated upon with the usual result—little or no improvement and only too frequently the death of the patient. Naturally, the condition is a most serious one and even the most dangerous operative procedures can be considered; no operation, however, should be advised to drain the ventricles unless it is definitely known that the ventricles are blocked and that the condition is not one of hydrocephalus externa. This fact has been so frequently overlooked that I feel that many needless and most tremendous operations have been preformed for the condition of hydrocephalus interna, which was not present—merely a hydrocephalus externa.

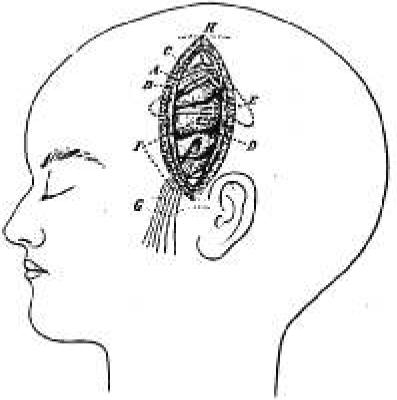

FIG. 1.—Diagrammatic sketch showing, through retracted temporal muscle (*A*), temporal fascia (*B*), and opened dura (*C*), the underlying cerebral convolutions and the Sylvian fissure (*D*). The dotted triangle (*E*) represents the lateral ventricle with the six linen strands (*F*) extending out from the ventricle through the upper temporal convolution beneath the Sylvain fissure. The scalp incision extends from the zygomatic arch (*G*) to the parietal crest (*H*).

If we exclude those cases of internal hydrocephalus due to a tumor or cyst formation in the posterior midbrain and subtentorially producing a direct pressure blockage of the ventricles it is my belief that practically all of the cases of hydrocephalus are the result of a diffuse condition—an original meningitis in its various forms; if its consequent adhesions and exudative thickenings are so situated as to block the exit of the cerebrospinal fluid into the subarachnoid spaces either in the aqueduct of Sylvius or at the foramina of Majendie and Luschka then we shall have produced the internal type of hydrocephalus; however, if the resulting adhesions, thickenings, etc., are not so situated as to block the ventricles, an internal hydrocephalus naturally does not occur, but still we shall have formed the external type of hydrocephalus because the cerebrospinal fluid cannot escape from the subarachnoid spaces of the cerebrospinal canal through its veins, sinuses, possibly lymphatics, etc., on account of their thickened walls and blocked stomata of exit due to the former meningitis of varying degree. Naturally, no operative procedure merely designed to drain the ventricles into the subarachnoid spaces would be of any definite benefit in these cases; even if the openings of such drainage tubes did not themselves become blocked (and it is rare for them not to become blocked by adhesions, surrounding tissues, etc.), we should merely have the condition of internal hydrocephalus changed to one of external hydrocephalus, which is, to be sure, possibly a less serious condition in that the cerebral cortex is not so compressed and thinned as results from ventricular dilatation; besides, some cerebrospinal fluid may now escape through channels of exit only partially obstructed, and so a slight improvement is possible; however, unless the condition of external hydrocephalus is relieved so that the cerebrospinal fluid

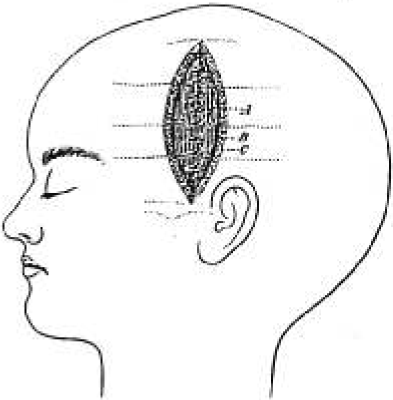

FIG. 2.—The six linen strands are brought out through the sutured temporal muscle (*A*) and temporal fascia (*B*) (to be sutured), and their ends are placed beneath the scalp (*C*) in the subcutaneous tissues—rich in lymphatics.

can escape from the subarachnoid spaces and the cerebrospinal canal then the end result is merely delayed; a normal child is impossible.

It is interesting to note that epilepsy occurs more frequently in the external type of hydrocephalus than in the internal type; this may be due to the greater cortical irritation of this supracortical blanket of cerebrospinal fluid in these cases and also to the possibly greater corticomeningeal damage at the time of the original meningitis. Again, the great frequency of spina bifida associated with hydrocephalus merely illustrates the etiological factor in their production, a meningitic blockage of the stomata of exit in the walls of the veins, sinuses, possibly lymphatics, etc., so that the cerebrospinal fluid cannot escape from the spinal canal;[3] therefore an increased intradural pressure results in a protrusion and hernial sac formation in the lumbar region, the weakest and the latest region of the spinal closure; the fact that spina bifida occurs in about 35 per cent. of the cases of hydrocephalus, and that it is almost never associated with the internal type of hydrocephalus merely emphasizes the method of its production—that is, an early meningeal inflammation with a general blockage of the cerebrospinal fluid causing an increase of the intradural pressure so that the hernial protrusion of a spina bifida results with or without an external hydrocephalus; it is most rare, however, for a spina bifida to occur without there being definite signs of a mild external hydrocephalus.

Again, it has frequently been observed following the repair of a spina bifida, especially within a few days after birth, that a marked enlargement of the head occurred in a large number of the patients; this condition of external hydrocephalus might have resulted whether the spina bifida had been operated upon or not; but it does seem that the operative repair of the spina bifida by the removal of the large hernial sac, might have lessened the excretory surface of the cerebrospinal canal to such an extent that an increased amount of cerebrospinal fluid collected sufficient to cause the head to enlarge; it should be stated, also, that in these children the early operative removal of the spina bifida protrusion is usually performed when the hernial sac is enlarging, and thus the safety valve, as it were, of the increased intradural pressure, due to a blockage of the cerebrospinal fluid, is removed, and therefore the head, the next weakest portion of the cerebrospinal canal, will enlarge unless the fluid can be successfully drained.

In three patients I have been obliged to use the cranial method of drainage described in this paper after the spina bifida had been repaired. However, in this series of 41 patients an external hydrocephalus appeared several weeks after birth in 9 patients having the condition of spina bifida which had not been operated upon;

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[3] Sharpe, Norman: Spina Bifida; an Experimental and Clinical Study, Ann. Surg., February, 1915, pp. 151–165.

in these cases it would seem that the spina bifida relief of the increased intradural pressure was not sufficient to prevent an enlargement of the head.

In this series of 41 operated patients we have used the same method of drainage, except in the first 2 cases, upon whom merely a bilateral subtemporal decompression was performed. Thirteen of the patients died, all of them with the exception of one child within thirty-six hours after the operation; that is, they could not survive the operative shock due to the sudden loss of a large amount of cerebrospinal fluid (this loss is now controlled and greatly lessened by elevating the head of the patient at the time of the ventricular opening). These patients were of the extreme type, very much emaciated, owing to their difficulty in swallowing, and were therefore bad operative risks. Of the remaining 28 patients upon whom the operation has been performed the results have been most encouraging. Their ages ranged from ten days to four years; naturally, the younger the child the less damaged is the cerebral cortex by the increased intracranial pressure; besides, the younger children stand the operative procedure as well and possibly better than the older ones, due most probably to the fact that many of the younger children are in a better physical condition than the ones subjected to a high intracranial pressure for a longer period of time; also to the fact that the cortical cells are not so highly developed in the younger children, and therefore they are apparently less upset by the operative procedure.

The object of the operation has been to drain permanently the ventricles (in the internal type of hydrocephalus) and the subarachnoid and subdural spaces (in the external type of hydrocephalus) outward beyond the cerebrospinal canal, that is, beyond the dura into the subcutaneous tissues of the scalp, a most absorptive area being well supplied with lymphatics. As drainage tubes buried in the body tissues always become blocked by adhesions, connective tissue, and surrounding tissues, etc., six linen strands are therefore inserted into the ventricles in the internal type and merely into the subarachnoid and subdural spaces in the external type of hydrocephalus and their ends brought out through the temporal muscle and temporal fascia beneath the scalp in a stellate manner; as these linen strands cannot be absorbed in the body tissues until four to six months have elapsed it is hoped that by the time of their absorption these artificial channels would be lined with endothelium or epithelium and thus their permanency and patency be assured.

In the laboratory the experimental production of internal hydrocephalus in puppies of ten days and two weeks of age has been performed by the method described by Dandy and Blackfan; that is, the aqueduct of Sylvius was blocked by a cotton plug through a suboccipital exposure; within twelve hours the ophthalmoscope revealed choked disks of varying degree showing high intracranial pressure due to a blockage of the ventricles; unless this condition was relieved a secondary optic atrophy would result (as was permitted to occur in one dog so that it apparently became blind.)[4] In the other dogs which survived the first operation of ventricular blockage a subtemporal decompression was now performed (in three dogs a bilateral opening was made), and the linen strands inserted into the lateral ventricle and allowed to extend outward through the cortex, the opened dura, through the temporal muscle and temporal fascia into the subcutaneous tissues of the scalp. It was most interesting to observe a gradual subsidence of the choked disks in all but three of these nine dogs; in none of them, however, did the eye-grounds become as normal as before the operation, there being always an edematous blurring of the optic disks showing that the relief of the ventricular blockage had not been entirely complete. These dogs all died (three of them as a result of the internal hydrocephalus), within ten weeks after the drainage of the ventricles with the linen strands, which had therefore not yet been absorbed; so that it was still not ascertained whether the artificial canals formed by the linen strands would have remained as permanent patent channels of drainage for the cerebrospinal fluid or not.

However, one of the children having the condition of external hydrocephalus upon whom I had performed a right ventricular drainage on June 19, 1916, and then a left ventricular drainage on July 7, 1916, this child after making a definite improvement and three months after the first drainage operation, fell out of a high chair at home on September 9, 1916, fracturing the base of the skull. It was brought to the Polyclinic Hospital upon my service and died on October 13, 1916. An examination of the brain revealed an almost complete absorption of the linen strands used at the first operation, whereas the strands of the second operation were only slight absorbed. The report of the pathologist, Dr. F. M. Jeffries, regarding the condition of the artificial canals in the temporal muscle and fascia and in the subcutaneous tissues of the scalp showed that the linen strands brought out through the temporal muscle and temporal fascia at the first operation were almost entirely absorbed, being surrounded by numerous giant cells embedded in proliferative structures rich in bloodvessels and exudative cells. As this was a case of external hydrocephalus no linen strands had been inserted in the ventricles; it will therefore be necessary for us to wait until an examination of such a brain can be made—that is, a case of internal hydrocephalus—and at least six months after the linen strands have been placed in the ventricle in order to ascertain whether their artificial canals will remain as permanent patent channels of exit for the cerebrospinal fluid by being lined with endothelial cells or not. The fact, however, that the children operated upon have improved and are still improving is of such significance to warrant the assumption that at least the artificial canals in the cerebral cortex are still patent.

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[4] This experimental work will be reported in detail later.

With the exception of 6 of the total 28 children living after this operation of drainage, all of them have improved and are still improving; 5 of them have improved mentally and physically in every way, all of these children having been operated within one year after birth; 7 of the patients unable to walk before the operation are now walking, although with difficulty. The different stages of papillo-edema and choking of the optic disks producing a secondary optic atrophy and the visual impairment in these patients have been improved in each case unless a complete secondary optic atrophy had already occured.

Naturally, sufficient time has not yet elapsed to permit an opinion regarding the permanency of this improvement, and whether any of the younger patients will be normal and remain normal: but I do feel that the operative method here used does offer a chance of improvement to a large number of patients formerly considered hopeless; if a definite improvement can be obtained in the older children, then surely an earlier operation in the younger children when the condition is first observed and before the brain has been permanently damaged, should make a more hopeful prognosis possible. I am deeply 'ndebted to Drs. McHenry and Hunt for their assistance; also to Dr. Lott for the apparatus devised by him to insert the linen strands into the ventricle; a description of it and the technic will be described in detail later.

EFFECT OF NUCLEIN INJECTION UPON THE LEUKOCYTES OF DOGS.

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The result of work upon the effect of nuclein in experimental animals has not been uniform, probably because under this name both the acid and its sodium salt have been employed and no standardization has been attempted. In approaching this subject, we determined to employ the product accepted by the American